Fetal Urinary Tract Abnormalities

24th Annual Ob-Gyn Ultrasound Update
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Objectives

- Normal Fetal Urinary Tract
- Abnormals
  - Agenesis
  - Ectopic
  - Obstructive
  - Dysplasia
Normal GU Anatomy

• **Kidneys**
  – Timeline
    • 10-12 weeks: Visible
    • Size directly proportional to GA
    • 3\textsuperscript{rd} trimester: Pyramids and cortex visibly separate

  – *Ultrasound appearance:*
    • Posterior, mid-abdomen
    • Flanks spine
    • Transverse, Sagittal, Coronal views
    • Capped by adrenals
Normal GU Anatomy

• Kidneys

![Graph showing kidney length vs. gestational age](image-url)
Normal GU Anatomy

• Kidneys @ 12 weeks
Normal GU Anatomy

- Kidneys @ 16 weeks

http://www.fetalultrasound.com/online/text/3-016.htm
Normal GU Anatomy

- Kidneys @ 20 weeks
Normal GU Anatomy

- Kidneys @ 28 weeks
Normal GU Anatomy

- Renal arteries
Normal GU Anatomy

• **Bladder**
  – Can be seen in 1st trimester

  – Ultrasound appearance:
    • Round/rectangular anechoic fluid-filled space in pelvis
    • Flanked by umbilical arteries
Normal GU Anatomy

- Bladder & 3VC
Normal GU Anatomy

- **Ureters**
  - Should not be visible in normal anatomy

- **Urethra**
  - Should not be visible in normal anatomy
Urinary Tract Abnormalities
Abnormal: Renal Agenesis

• Bilateral
  – 1-3 per 10,000
  – Lethal
  – Sporadic/isolated or part of syndrome
  – Recurrence risk: 3.5-5.9%

  – Ultrasound appearance:
    • Kidneys not identifiable
    • Color Dopplers do not reveal renal arteries (PPV 82%)
    • Adrenals “laying down”
    • Empty bladder (on prolonged or repeat examination)
    • Oligohydramnios after 16 weeks
Abnormal: Renal Agenesis
Abnormal: Renal Agenesis

• **Unilateral**
  – 3-4x more common than bilateral
  – Good prognosis

  – *Ultrasound appearance:*
    • Compensatory hypertrophy: AP:TR diameter ratio
      – Normal 2\textsuperscript{nd} trimester: 0.84 (0.72-0.89)
      – Normal 3\textsuperscript{rd} trimester: 0.81 (0.65-0.89)
      – Unilateral agenesis: \(\geq 0.9\) (high sensitivity/specificity)
    • Normal bladder
    • Normal AFV
Abnormal: Renal Agenesis
Abnormal: Renal Agenesis

- “Potter”
  - Edith Louise Potter, 1901-1993
  - Classic Potter Syndrome
    - Bilateral renal agenesis
  - Sequence
    - “Oligohydramnios sequence”
    - Physical appearance of a fetus/neonate due to persistent oligohydramnios
    - Clubbed feet, contractures, pulmonary hypoplasia, cranial anomalies, low-set ears, flattened nose/face, micrognathia, IUGR
Abnormal: Renal Agenesis

• “Potter”

  – Ultrasound findings
    • LIMBS: Clubbed feet, contractures
    • THORAX: pulmonary hypoplasia
    • HEAD/NECK:
      – Cranial abnormalities
      – Low-set ears
      – Flattened nose/face
      – Micrognathia
      – IUGR
Abnormal: Ectopic Kidney

- 1:7000 births
- Locations:
  - Pelvis: most common
  - Crossed renal ectopia
Abnormal: Ectopic Kidney

• Pelvic
Abnormal: Ectopic Kidney

- Pelvic

Fusion point of Lt + Rt kidneys

Crossed fused renal ectopia
Abnormal: Ectopic Kidney

- Thoracic
Abnormal: Obstructive

- **Obstructive uropathies**
  - Portion or entire urinary tract
  - Terminology
    - Hydronephrosis: abnormal dilation of renal pelvis and calyces
    - Pyelectasis: abnormal dilation of pelvis only
  - Sites:
    - Ureteropelvic junction (most common)
    - Urethra
    - Ureterovesical junction
  - Male > Female (3:1)
Abnormal: Obstructive

• Pyelectasis / Hydronephrosis
Abnormal: Obstructive

- Pyelectasis
  - Demographics
    - Male > Female (3:1)
    - Left > Right
  - GA dependent

<table>
<thead>
<tr>
<th>GA</th>
<th>Normal</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
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<tbody>
<tr>
<td>15-20 w</td>
<td>&lt;4 mm</td>
<td>4-7 mm</td>
<td>&gt;7 mm</td>
<td></td>
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<tr>
<td>20-30 w</td>
<td>&lt;5 mm</td>
<td>5-8 mm</td>
<td>9-15 mm</td>
<td>&gt; 15 mm</td>
</tr>
<tr>
<td>&gt;30 w</td>
<td>&lt; 7 mm</td>
<td>7-9 mm</td>
<td>10-15 mm</td>
<td>&gt;16 mm</td>
</tr>
</tbody>
</table>

- Outcomes similar between sexes

Mandel, 1990; Society of Fetal Urology
Abnormal: Obstructive

- Pyelectasis / Hydronephrosis Outcomes

- 11,465 women @ 18-23 weeks
- 2.3% with hydronephrosis
  - 80.6% Mild
    - None required surgery
  - 19.4% Mod-Severe
    - 1 in 3 required surgery
Abnormal: Obstructive

• Pyelectasis / Hydronephrosis Evaluation
  – Association with T21
    • 9.1% of T21 with some pyelectasis
    • Isolated pyelectasis = LR 1.5-3.8
    • Pyelectasis + other anomalies = LR 19.2
    • Compared to all other organ systems, lowest correlation with T21
  – Recurrence risk
    • RR 6.1
Abnormal: Obstructive

- Pyelectasis / Hydronephrosis
Abnormal: Obstructive

- Pyelectasis / Hydronephrosis
Abnormal: **Obstructive**

- **Pyelectasis / Hydronephrosis**
Abnormal: Obstructive

- Pyelectasis / Hydronephrosis
  Postnatal follow-up

  - US @ Birth
    - If severe, VCUG at 2-4 weeks
  
  - US @ 1 month

  - Serial US and UTI surveillance q 6-12 months
Abnormal: Obstructive

- **Ureteropelvic Junction Obstruction (UPJO)**
Abnormal: Obstructive

• UPJO
  – Most common cause of neonatal hydronephrosis
  – 50% of congenital urinary tract abnormalities
  – Cause: Kinks, bands, fibrous adhesions, ureteral valves, abnormal insertion, unusual shapes of pyeloureteral outlet
  – 10-30% bilateral
  – M>F (5:1)
  – Sporadic, but may be familial
Abnormal: Obstructive

• **UPJO**
  - Associated anomalies:
    • MCDK
    • Cardiovascular
    • NTD
    • Hirschsprung
    • Imperforate anus
    • Esophageal atresia
  - Amniotic fluid
    • Oligohydramnios if severe and bilateral
    • Paradoxical polyhydramnios
Abnormal: Obstructive

- UPJO
Abnormal: Obstructive

• UPJO
Abnormal: Obstructive

- Ureterovesical Junction Obstruction (UVJO)
Abnormal: Obstructive

- **UVJO**
  - “Megaureter” or “Hydroureter”
  - Causes: Ureteral stenosis, fibrosis, abnormal muscularis, external compression, duplicated collecting system, ectopic ureterocele
  - M>F
  - Sporadic, but may be familial

- Ultrasound appearance:
  - Tortuous, anechoic, tubular structure traced from origin in renal pelvis to urinary bladder
Abnormal: Obstructive

- Hydroureter
Abnormal: Obstructive

- UVJO
  - Associated anomalies:
    - Ureterocele
    - Contralateral agenesis
    - Multicystic kidney disease
    - Hirschsprung disease
Abnormal: Obstructive

- Ureterocele

https://iame.com/online/fetal_uropathy/content.php
Abnormal: Obstructive

- Posterior Urethral Valves (PUV)
Abnormal: Obstructive

• **PUV**
  – Most common cause of distal urinary tract obstruction

  – *Ultrasound findings:*
    • Marked distension & hypertrophy of bladder
    • Keyhole sign
    • ± Hydronephrosis & hydroureter (bilateral)
    • ± Oligohydramnios
    • ± Renal dysplasia
Abnormal: Obstructive

- PUV

- *Megacystis:*
  - **Normal:** Bladder length > 7 mm before < 10 weeks
  - **Intermediate:** Bladder length at 7-15 mm
    - Spontaneous resolution in 90% by 20 weeks
  - **Severe:** Bladder length > 15 mm
    - Progressive obstructive uropathy
Abnormal: Obstructive

- PUV
Abnormal: Obstructive

• PUV
Abnormal: Obstructive

• PUV
Abnormal: Obstructive

• PUV
Abnormal: Obstructive

- PUV
Abnormal: Obstructive

• PUV
  – Associated anomalies (20-25%)
    • Cardiovascular anomalies
    • Tracheal hypoplasia
    • Scoliosis
    • Imperforate anus
Abnormal: Obstructive

- Urethral atresia
  - Ultrasound findings:
    - Similar to posterior urethral valve initially
    - Anhydramnios
    - Pulmonary hypoplasia
Abnormal: **Cystic Disease**

- **Potter Types**
  - *Type I*:
    - Autosomal recessive (infantile) polycystic kidney disease (AR-PKD)
  - *Type II*:
    - MCDK disease
  - *Type III*:
    - Autosomal dominant (adult) polycystic kidney disease (AD-PKD)
  - *Type IV*:
    - Renal dysplasia

Dighe, et al. 2011
Abnormal: Cystic Disease

• Potter Types
  – *Type I*: AR-PKD
    • Chromosome 6p
    • Abnormal collecting tubules
    • Associated anomalies:
      – Hepatic cysts
      – Biliary duct hyperplasia
      – Portal hypertension

Dighe, et al. 2011
Abnormal: Cystic Disease

- Potter Types
  - *Type I*: AR-PKD
  - Types
    - Perinatal: Most common
      - Renal failure in-utero
      - Stillbirth or Neonatal death
    - Neonatal
    - Infantile
    - Juvenile

Dighe, et al. 2011
Abnormal: Cystic Disease

- **Potter Types**
  - *Type I:* AR-PKD

  - *Ultrasound findings*
    - 1-2 mm cysts at periphery
    - Renal pelvis, calyces, ureters normal
    - Bilaterally enlarged kidneys
    - Hyperechoic
    - Oligohydramnios
    - Small bladder

Dighe, et al. 2011
Abnormal: Cystic Disease

- Potter Types
  - *Type I*: AR-PKD

Dighe, et al. 2011
Abnormal: Cystic Disease

• Potter Types
  – Type II: MCDK
    • Most common type leading to ESRD in children
    • Sporadic
    • Laterality
      – Bilateral: 20%
      – Unilateral: 80% with contralateral anomalies in 40%
      – Segmental
  • Subcategories
    – IIA = Large kidneys
    – IIB = Small or normal kidneys

Dighe, et al. 2011
Abnormal: Cystic Disease

- Potter Types
  - *Type II*: MCDK

**Ultrasound findings:**
- No normal parenchyma
- Normal, small, or large size
- Cysts = Dilated collecting tubules
- No communication between enlarged tubules
- Renal artery small or absent

Dighe, et al. 2011
Abnormal: Cystic Disease

- Potter Types
  - Type II: MCDK
Abnormal: Cystic Disease

• Potter Types
  – Type II: MCDK
    • Meckel-Gruber
Abnormal: Cystic Disease

• Potter Types
  – *Type II*: MCDK
    • Associated anomalies:
      – GU: Atretic ureter, Pelvoinfundibular atresia,
      – CV: Cardiovascular anomalies
      – GI anomalies, CDH
      – Face: Cleft
      – MSk: absence of radius, thumbs
      – NTD, microcephaly
      – Meckel-Gruber syndrome (AR)
      – Apert syndrome (AD)

• Hypertension

Dighe, et al. 2011
Abnormal: Cystic Disease

- **Potter Types**
  - *Type III: AD-PKD*
    - Most common form of hereditary renal cystic disease
    - PKD1 gene (chr 16p) and PKD2 gene (4q)
    - Can be present in fetal or neonatal period
  - **Ultrasound findings:**
    - Symmetrically enlarged
    - Echogenic
    - Small cysts
    - Bladder & AFV normal
  - Associated anomalies: Cysts elsewhere (liver, pancreas, spleen, CNS)
  - Family History; US of parents’ kidneys

Dighe, et al. 2011
Abnormal: Cystic Disease

• Potter Types
  – Type IV: Cystic Renal Dysplasia
    • Sequelae of obstructive uropathy
    • Degree of dysplasia correlates with obstruction
    • Hard to distinguish from MCDK
    • Poor prognostic factors:
      – Urinomas
      – Ascites
      – Oligohydramnios

Dighe, et al. 2011; Mandel, 1990; Society of Fetal Urology
Abnormal: Dysplasia

- Potter Types
  - Type IV: Cystic Renal Dysplasia

- Ultrasound findings predictive of dysplasia

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<th>Sens</th>
<th>Spec</th>
<th>PPV</th>
<th>NPV</th>
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<tr>
<td>Cortical cysts</td>
<td>44%</td>
<td>100%</td>
<td>100%</td>
<td>56%</td>
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<tr>
<td>Echogenic kidneys</td>
<td>74%</td>
<td>80%</td>
<td>89%</td>
<td>57%</td>
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**Abnormal: Dysplasia**

- **Potter Types**
  - *Type IV: Cystic Renal Dysplasia*
  - **Biochemical findings**

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<th>Normal</th>
<th>Abnormal</th>
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<tr>
<td>Na⁺</td>
<td>$&lt;100$ mEq/L</td>
<td>$&gt;100$</td>
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<tr>
<td>Cl⁻</td>
<td>$&lt;90$ mEq/L</td>
<td>$&gt;90$</td>
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<tr>
<td>Ca++</td>
<td>$8$ mg/dL</td>
<td>$&gt;8$</td>
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<tr>
<td>Osm</td>
<td>200-210 mOsm/L</td>
<td>$&gt;200-210$</td>
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<tr>
<td>B₂-microglobulin</td>
<td>$&lt;4$ mg/dL</td>
<td>$&gt;6-10$</td>
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<tr>
<td>Protein</td>
<td>$&lt;40$ mg/dL</td>
<td>$&gt;40$ mg/dL</td>
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Mandel, 1990; Society of Fetal Urology
Conclusions

- Understand normal embryology and fetal development
- Evaluate entire urinary tract
- Look for associated anomalies
- Genetics consultation and family history
Questions?